

Inherited Heart Disease in the Dog

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Several forms of cardiac disease in the dog have now been demonstrated to be inherited or "familial." These include both congenital and adult onset (often termed acquired) heart disease. It is essential to understand the differences in the terminology. The term congenital refers to defects present at the time of birth, generally developmental defects. Inherited defects are those that an animal is genetically programmed to develop. These defects may be present at birth (congenital) or may not become apparent until the animal is an adult (adult onset). Not all congenital defects are inherited as vice versa.

The most common inherited heart diseases in the dog include the congenital defects, subvalvular aortic stenosis, pulmonic stenosis and patent ductus arteriosus (PDA). The most common adult onset inherited diseases include dilated cardiomyopathy and endocardiosis. There are other inherited heart defects in dogs that are occasionally observed, however given the limited time allowed here, they will not be discussed in this session.

SUBVALVULAR AORTIC STENOSIS

Subvalvular aortic stenosis is a congenital heart defect characterized by a fibrous ridge or ring below the aortic valve. It is often not evident at birth, but can become more obvious as the puppy gets older, generally leveling off by 1 year of age. Dogs with mild disease may live a normal life span; however dogs with more moderate or severe disease can have a significantly shorter life span and die suddenly at less than 2 years of age, or develop congestive heart failure. A small percentage of affected dogs develop an infection on the heart valve (bacterial endocarditis).¹

Subvalvular aortic stenosis (SAS) is most commonly reported in the Newfoundland, Golden Retriever, Boxer and Rottweiler.² Although inheritance is suspected in several breeds it has only been proven in the Newfoundland.³ The inheritance pattern is not well understood, but appears to be inherited as autosomal dominant with variable penetrance in the Newfoundland. Inheritance is strongly suspected in other breeds.

The diagnosis of SAS in affected puppies is first suspected when a heart murmur is heard at the left base of the heart with a stethoscope by a veterinarian. Murmurs are fairly nonspecific sounds that suggest that a dog has heart disease. In many cases, the murmur may be detected at 8 weeks, but in some cases, it is not easily heard until they are a bit older (12-16 weeks). Further evaluation with a cardiac ultrasound (echocardiogram, echo) is needed to confirm the diagnosis since sometimes these murmurs can be "innocent" (developmental) or caused by a different defect.

Many owners who have heard of the inheritable nature of SAS in the dog have interest in screening their puppies. Although an initial screening can be performed by auscultation by a veterinarian, probably all dogs should have an echocardiogram performed prior to breeding at 12 months of age. Since the defect can progress slightly over the first year of life, it is difficult to accurately diagnose the final severity before 12 months of age. Doppler echocardiography is a technique that is highly user dependent. Therefore, an individual that is not properly trained in echocardiography may over- or under-diagnose the disease. We generally recommend that veterinary cardiologists perform the screening whenever possible, to avoid misdiagnosis. Affected dogs should not be used for breeding.

DILATED CARDIOMYOPATHY

The most commonly reported inherited adult onset canine heart disease is dilated cardiomyopathy (DCM).² Dilated cardiomyopathy is an adult onset, primary heart muscle disease. The onset of DCM results in the development of a dilated left ventricle with decreased systolic (pumping) function. Eventually, this may lead to increased pressure in

the chamber above the ventricle, the atria. Increased atrial pressure can result in the development of fluid in the lungs called pulmonary edema (congestive heart failure) which may make it difficult for the dog to breathe properly. Some dogs develop an abnormality in the electrical system of the heart (arrhythmia) which results in abnormal heart beats; this may result in sudden death. The development of clinical signs (heart failure, sudden death) may vary depending on the affected breed of dogs.

The diagnosis of DCM in affected dogs is first suspected when a heart murmur or gallop is heard with a stethoscope by a veterinarian. Murmurs and gallops are fairly nonspecific sounds that **suggest** that a dog may have heart disease. Other signs may include the development of a persistent cough (Doberman Pinschers), increased fluid in the abdomen (Great Danes) and fainting episodes (Boxers, Doberman Pinschers). Further evaluation with a cardiac ultrasound (echocardiogram, echo) should be performed as soon as possible to confirm the diagnosis.

Many owners who have heard of the inheritable nature of DCM in the dog have interest in screening their dogs prior to breeding. It should be remembered that DCM is an "adult-onset" disease. This means that it may develop at some point in adulthood. Some dogs develop a severe form of disease as a very young adult (8-12 months). However most breeds of dogs develop it at a more mature age. Therefore, it is very important to evaluate a dog **every year** to accurately screen. Screening may include a physical examination, echocardiogram and Holter monitor (ambulatory electrocardiogram). The individual tests needed will depend on the individual breed. All of these tests are techniques that are highly user dependent. Therefore, an individual that is not properly trained in cardiology may over- or under- diagnose the disease. We generally recommend that veterinary cardiologists perform the screening whenever possible, to avoid misdiagnosis.

Doberman pinscher

Annual echocardiography and ambulatory electrocardiography (Holter monitoring) are believed to be the best predictors of early DCM in this breed.^{4,5} Criteria that are believed to be indicators of early disease include left ventricular size (left ventricular diastolic dimension > 4.6 cm, systolic dimension > 3.8 cm). These numbers are based on average sized dogs and may not be valid for very large dogs. Annual Holter monitoring has also been recommended to detect Doberman pinschers that may develop ventricular arrhythmias before ventricular dilation. Adult Doberman pinschers with greater than 50 ventricular premature complexes (VPCs) per 24 hours, or couplets or triplets are suspect for the development of DCM.⁵

GIANT BREED DILATED CARDIOMYOPATHY

Giant breed dilated cardiomyopathy is used to describe DCM in the Irish Wolfhound, Great Dane, Scottish Deerhound and Newfoundland dog, among others. A high percentage of affected dogs in these breeds present with the electrical abnormality called atrial fibrillation. In some cases, atrial fibrillation may develop before any other evidence of underlying myocardial disease (chamber enlargement or systolic (pumping) dysfunction). However, not all dogs with atrial fibrillation will develop DCM, but these dogs should be carefully followed for the development of DCM and perhaps should be held out of breeding programs

Occasional cases of familial disease in the Great Dane, Newfoundland and Irish wolfhound have been identified. In the Great Dane, it is most likely an X-linked disease.⁶ Sons of affected females are at high risk of developing the disease; daughters of affected fathers are likely to be silent carriers. Since it is adult onset, all dogs should be screened annually with echocardiography.

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

Historically, boxers that presented with fainting episodes and electrical abnormalities called ventricular arrhythmias were diagnosed with dilated cardiomyopathy. However, this disease appears to have more similarities with arrhythmogenic right ventricular cardiomyopathy (ARVC) in human beings than with true dilated cardiomyopathy. This disease is now frequently being referred to as boxer ARVC.

This is a familial, adult onset disease that appears to be inherited as an autosomal dominant trait. Since this disease appears to present as an electrical abnormality, any screening efforts should be based on annual Holter monitoring and possibly, annual echocardiography. Clear criteria for affected status are still being determined and day to day variability of arrhythmias exist, so owners should be encouraged to screen annually rather than put emphasis on a single Holter reading. Interpretation of the Holter results can sometimes be challenging because strict criteria for this diagnosis does not exist. However since it is unusual for a normal dog to have any VPCs in a 24-hour period, the observation of > 100 VPCs, or periods of couplets, triplets or runs of ventricular tachycardia are abnormal. Supraventricular premature complexes may be seen but not frequently.

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SPEAKER INFORMATION

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